



Amyloidosis and Kidney Disease

Proteins are important building blocks for all body parts, including muscles, bones, hair, and nails. Proteins circulate throughout the body in the blood and are normally harmless. Occasionally, cells produce abnormal proteins that can settle in body tissue, forming deposits and causing disease. When these deposits of abnormal proteins were first discovered, they were called *amyloid*, and the disease process *amyloidosis*.

In recent years, researchers have discovered that different kinds of proteins can form amyloid deposits and have identified several types of amyloidosis. Two of these types are closely related to kidney disease. In *primary amyloidosis*, abnormal protein production occurs as a first step and can lead to kidney disease. *Dialysis-related amyloidosis (DRA)*, on the other hand, is a result of kidney disease.

Primary Amyloidosis

Primary amyloidosis occurs when the body produces abnormal protein fibers, which join together to form amyloid deposits in different organs, including the kidneys, where they cause serious

damage. Injured kidneys can't function effectively and may be unable to remove urea and other wastes from the blood. Elevated levels of these waste products can also damage the heart, lungs, brain, and digestive system.

One common sign of amyloidosis is the presence of abnormally high amounts of protein in the urine, a condition known as *proteinuria*. Healthy kidneys prevent protein from entering the urine, so the presence of protein may be a sign that the kidneys aren't working properly. A physician who finds large amounts of protein in the urine may also perform a biopsy—take a small sample of tissue for examination under a microscope—to confirm amyloidosis.

No effective treatment has been found to reverse the effects of amyloidosis. Combination drug therapy with melphalan (a cancer drug) and prednisone (an anti-inflammatory steroid drug) may improve organ function and survival rates by interrupting the growth of cells that produce amyloid protein. These are the same drugs used in chemotherapy to treat certain cancers, and they may have serious side effects, such as nausea and vomiting, hair loss, and fatigue.



National Institute of Diabetes and Digestive and Kidney Diseases
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Dialysis-Related Amyloidosis

Normal kidneys filter excess proteins from the blood, thus preventing levels from getting too high. When the kidneys don't work properly, as in patients receiving dialysis, another type of protein called *beta-2-microglobulin* may build up in the blood. When this occurs, beta-2-microglobulin molecules may join together, like the links of a chain, forming a few very large molecules from many smaller ones. These large molecules can form deposits and eventually damage the surrounding tissues and cause great discomfort. This condition is called *dialysis-related amyloidosis (DRA)*.

DRA is relatively common in patients who have been on dialysis for more than 5 years, especially among the elderly. Dialysis membranes don't effectively remove the large, complex beta-2-microglobulin proteins from the bloodstream. As a result, blood levels become elevated, and deposits form in bone, joints, and tendons. DRA may result in pain, stiffness, and fluid in the joints. Patients with DRA may also develop hollow cavities, or cysts, in some of their bones; these may lead to unexpected bone fractures. Amyloid deposits may cause tears in ligaments and tendons (the tissue that connects the muscle to the bone). Most patients with these problems can be helped by surgical intervention.

Half of the people with DRA also develop a condition called carpal tunnel syndrome, which results from the unusual buildup of protein in the wrists. Patients with this disorder may experience numbness or tingling, sometimes associated with muscle weakness, in their fingers and hands. This is a treatable condition.



Amyloid may build up in the wrist and cause bone cysts or carpal tunnel syndrome.

Unfortunately, no cure for DRA has been found, although a successful kidney transplant may stop the disease from progressing. However, DRA has caught the attention of dialysis engineers who are attempting to develop membranes that can more efficiently remove beta-2-microglobulin from the blood.

For More Information

For more information, contact the following organizations:

For information on primary amyloidosis:

**National Arthritis and Musculoskeletal and
Skin Diseases Information Clearinghouse**

National Institutes of Health

1 AMS Circle

Bethesda, MD 20892-3675

Phone: 1-877-22NIAMS (1-877-226-4267)

or (301) 495-4484

TTY: (301) 565-2966

Fax: (301) 718-6366

Internet: www.nih.gov/niams/healthinfo

Amyloidosis Network International, Inc.

7118 Cole Creek Drive

Houston, TX 77092-1421

Phone: 1-888-AMYLOID or (713) 466-4351

Internet: <http://amyloidosis.org>

*For information on dialysis-related
amyloidosis:*

National Kidney Foundation, Inc.

30 East 33rd Street

New York, NY 10016

Phone: 1-800-622-9010 or (212) 889-2210

Fax: (212) 689-9261

Email: info@kidney.org

Internet: www.kidney.org

National Kidney and Urologic Diseases Information Clearinghouse

3 Information Way

Bethesda, MD 20892-3580

Phone: 1-800-891-5390 or (301) 654-4415

Fax: (301) 907-8906

Email: nkudic@info.niddk.nih.gov

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Publications produced by the clearinghouse are carefully reviewed by both NIDDK scientists and outside experts. This fact sheet was also reviewed by Glenn Chertow, M.D., University of California at San Francisco, and William J. Stone, M.D., Vanderbilt University.

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U.S. DEPARTMENT OF HEALTH
AND HUMAN SERVICES
National Institutes of Health

NIH Publication No. 01-4694
April 2001

